Pharming Group NV

(NYSE Euronext: PHARM)

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Investment highlights

- Ruconest™ (rhC1-INH) franchise currently launching in Europe
  - Indication: acute treatment of hereditary angioedema (HAE) attacks
  - Partnered with SOBI
- US Phase III (Rhucin®) program close to completing
  - Data expected in 2012
  - Partnered with Santarus
- Follow-on indications underway for unmet needs in larger markets:
  - Antibody Mediated Rejection (Kidney Transplant)
  - Ischemia Reperfusion Injury (Delayed Graft Function, Acute Myocardial Infarction)
- Technology platform poised to replicate rhC1-INH success
  - Low cost, scalable and validated platform
  - Ideally suited for production of complex proteins

Recent & Upcoming Milestones

- EU & US commercialisation deals signed ✓
- EU launch of Ruconest ✓
- Progress on US development pathway for Rhucin ✓
- **Continue EU rollout of Ruconest** ✓
- **Expand geographic coverage of Rhucin franchise** ✓
- **Expand rhC1-INH platform**
  - **Initiate reperfusion injury programmes**
- **Leverage potential of the platform**
  - **New proteins, new indications**

Pipeline

	Indication	R&D	Pre Clinical	Phase I	Phase II	Phase III	Registration	Market
Ruconest™ / Rhucin®								
Ruconest™ (rhC1INH) (Europe)	Hereditary Angioedema							
Rhucin® (rhC1INH) (US)	Hereditary Angioedema							
rhC1INH additional indications								
rhC1INH	Antibody Mediated Rejection (Kidney)							
rhC1INH	Delayed Graft Function (Kidney)							
rhC1INH	Acute Myocardial Infarction							
Legacy pipeline								
rhFibrinogen	Fibrinogen deficiency							
rhCollagen	Tissue repair							
hLactoferrin	Nutritional applications							

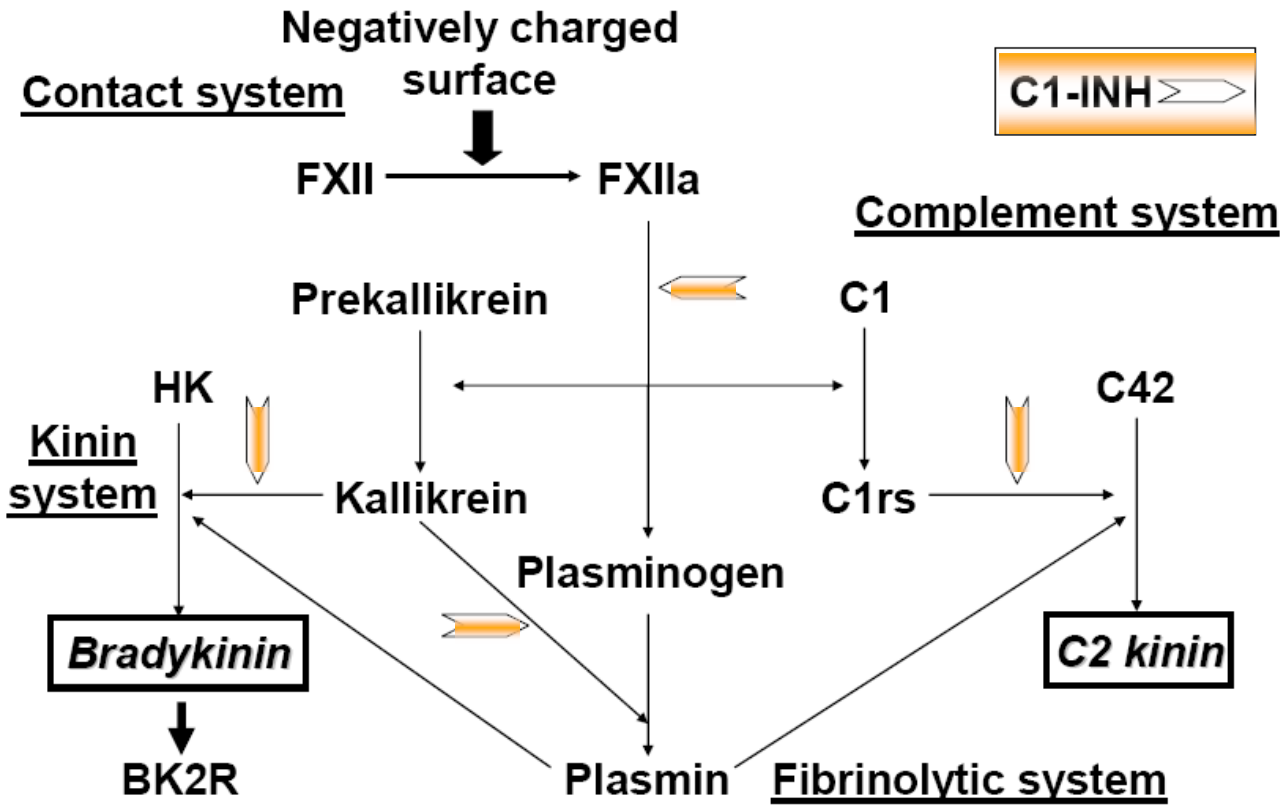
Core focus products/indications

Partnerships + risk sharing models for further development

HAE & C1 inhibition

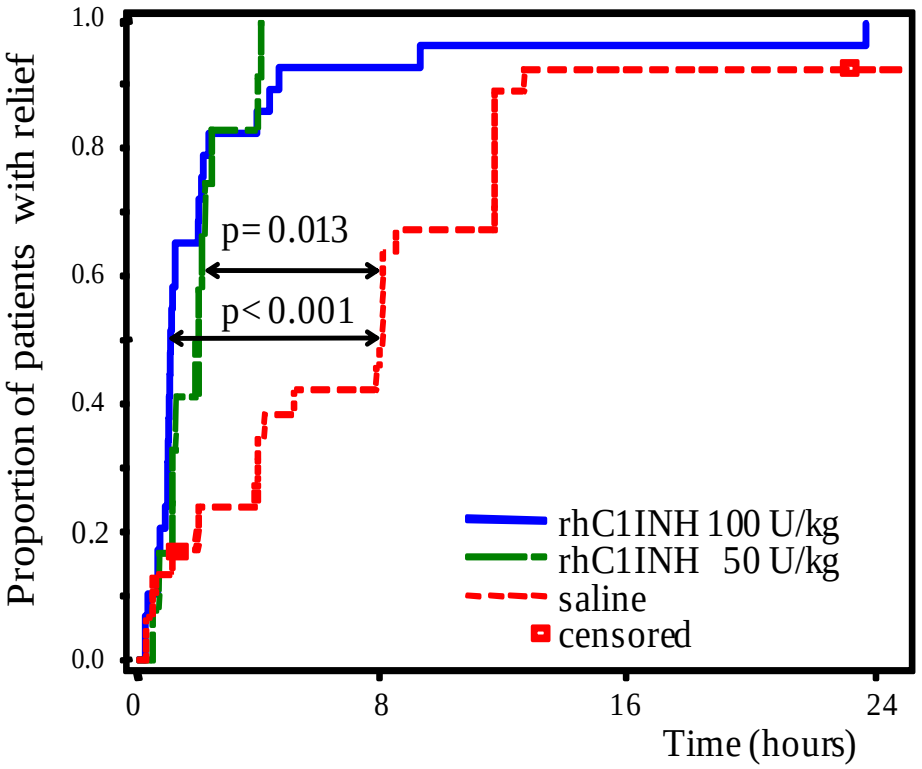
- Rare genetic disorder caused by mutations in the gene encoding C1 esterase inhibitor (C1-INH)
  - Low functional levels of the complement control plasma protein C1-INH
  - Patients present with swelling, severe abdominal pain, or acute airway obstruction
- Prevalence of disease estimated at 1 in 30,000
- 8+ swelling episodes requiring treatment per patient per year
  - Despite wide spread long term steroid prophylaxis
  - Laryngeal attacks are potentially lethal
  - Significant Quality of Life issues for patients given frequency of attacks
- Three systems involved in HAE (Complement, Contact, Fibrinolytic)
  - C1 inhibitor (missing protein) controls all three systems
- Treatment with C1 inhibitor considered ‘gold standard’ by clinicians

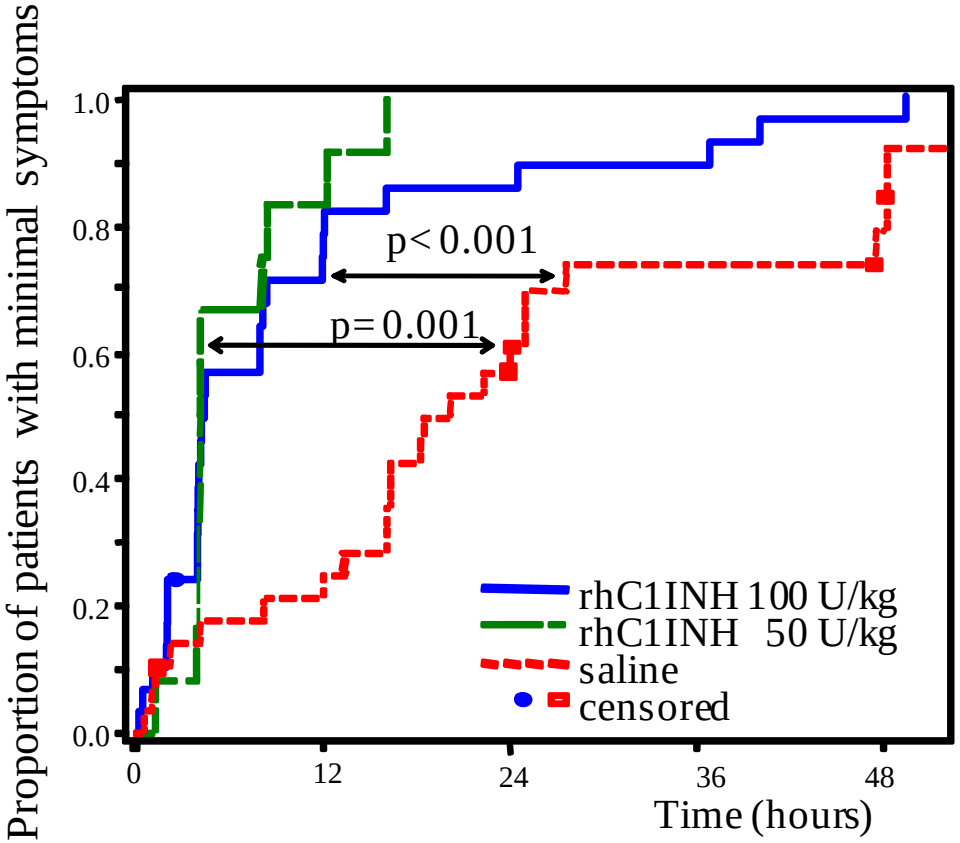
C1INH - function



Adapted from Caliezi et al 2000

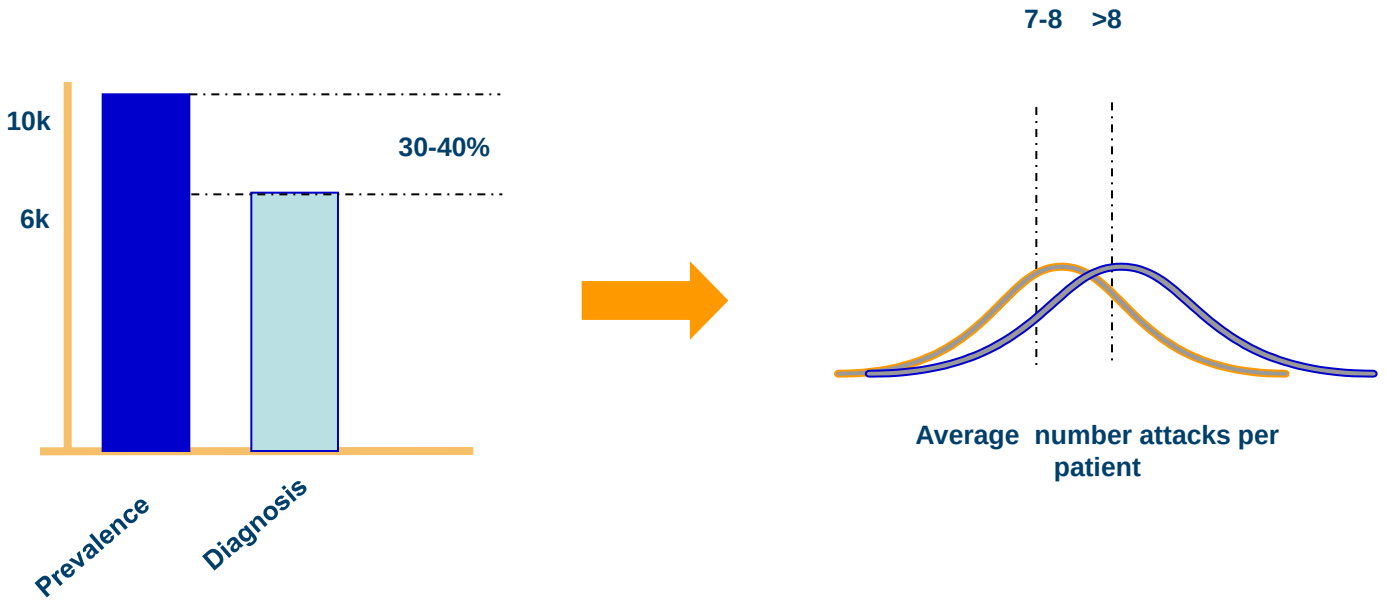
Clinical Efficacy Demonstrated in both RCTs





Zuraw, JACI 126:821 2010

Market Drivers: Patient numbers

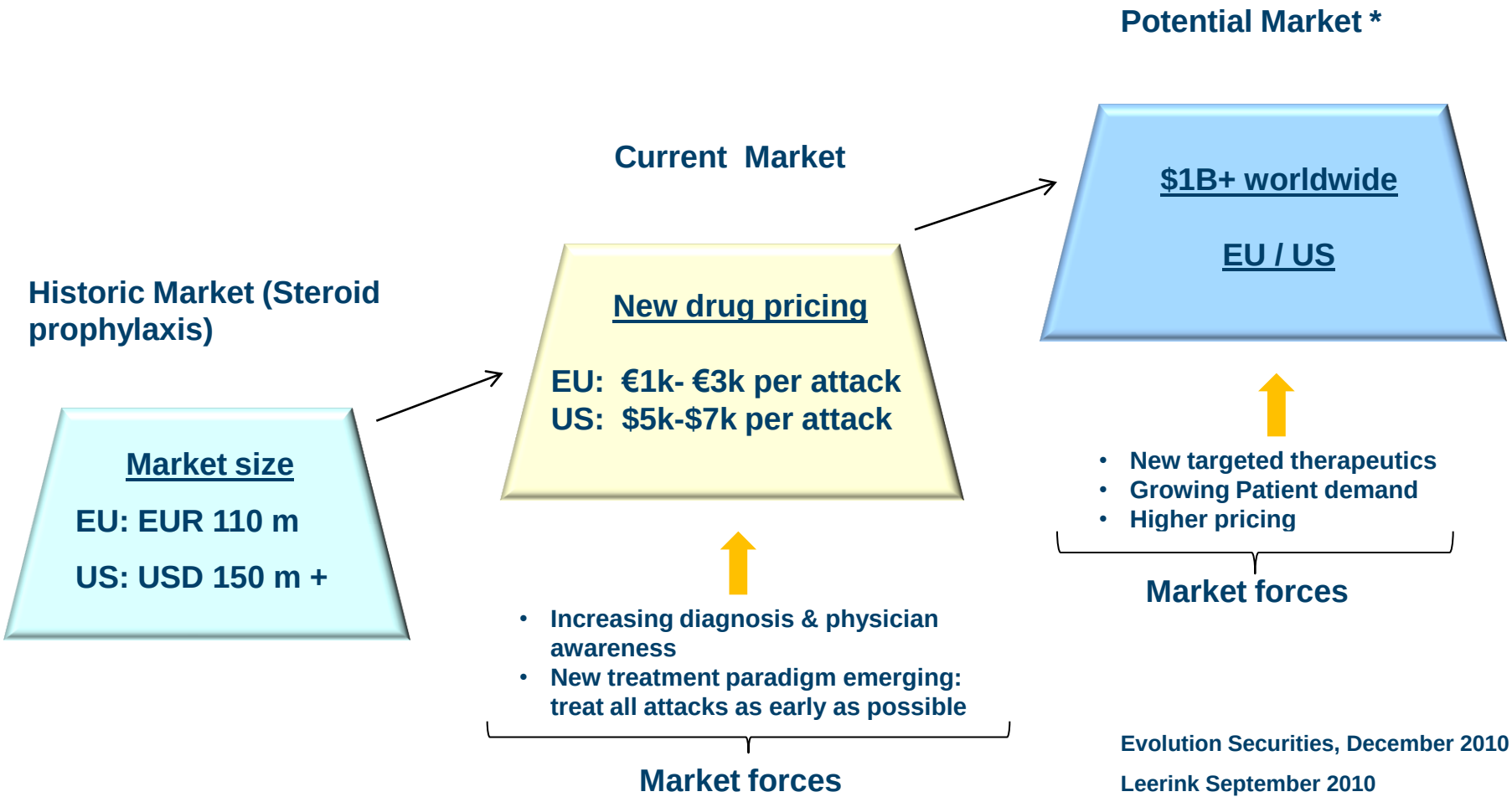


- There are a wide range of prevalence estimates: 1 in 10K–50K individuals
- 75% of patients present with symptoms before age 15
- Misdiagnosis is common

- Estimates suggest that the US & EU have ~10,000 patients, with 6,500 patients seeking treatment, each having ~8 moderate/severe attacks per year
- Pts have 1-2 attacks per month but seek treatment every other month

Changing Market Dynamics of HAE

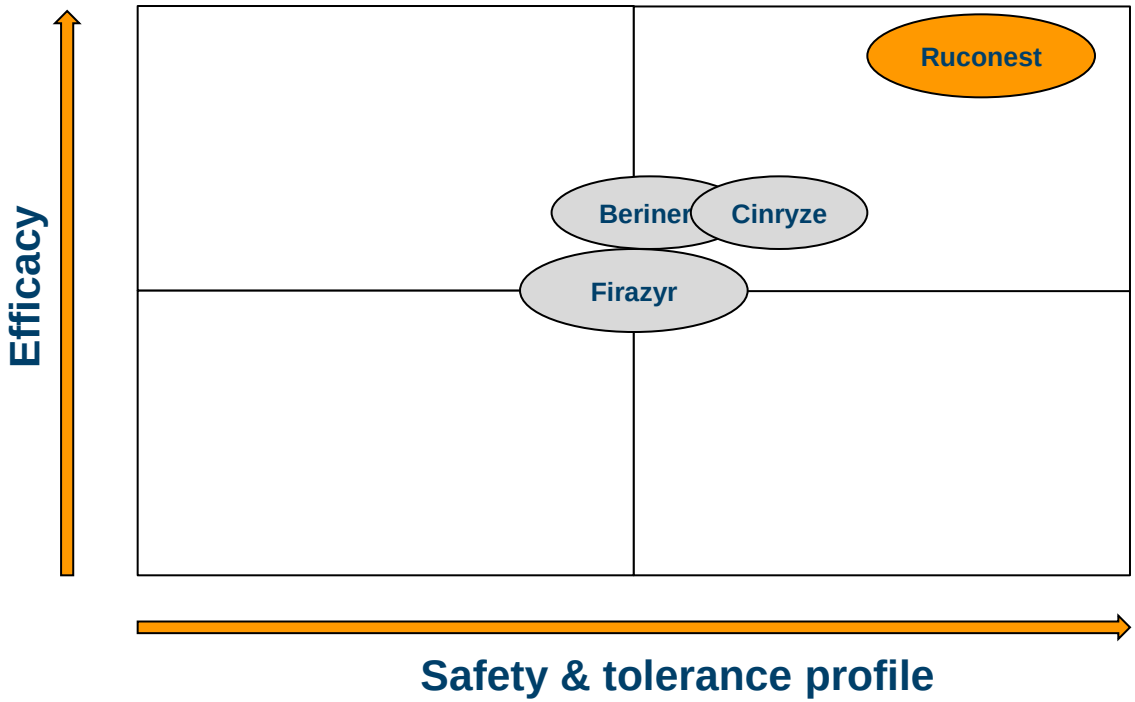
Transforming the market opportunity



Evolution Securities, December 2010

Leerink September 2010

Potential positioning: EU (Acute)



Berinert/ Cinryze:

- Potentially under-dosed (500-1500 U) absence of clinical effects published
- Significant level of impurities may explain side effect profiles

Firazyr:

- Almost 100% (very) painful SC injection
- Relatively high level of estimated attack recurrence (32%)

Craig et al: JACI; 2009/ Zuraw et al: NEJM 2010/ Icatibant Clinical Briefing Document, CDER, FDA, 2011./ Aberer, et al. Ann Allergy Asthma Immunol 2010; 105(5):P238

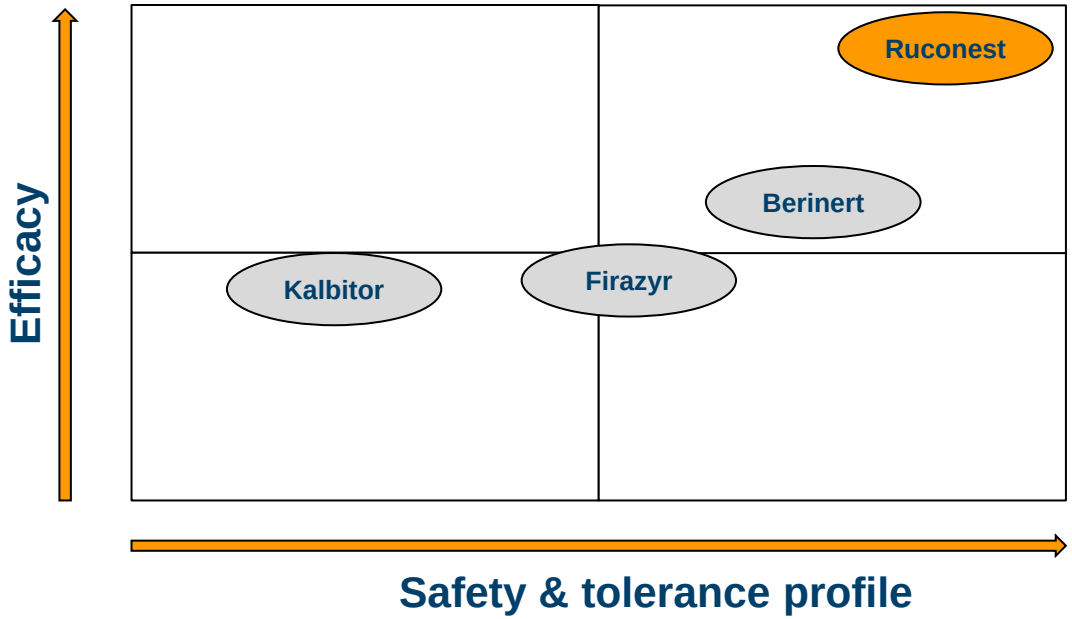
EU developments

- EU rollout continuing
  - Scandavia, Germany, UK, France & Netherlands
  - Q2, 2011 sales up 26% on Q1 to EUR 0.2 million
  - Reimbursement at national and local levels ongoing
- Increased geographical coverage
  - SOBI granted additional rights
    - Balkans, North Africa & Middle East
  - Former Esteve territories (Spain Portugal & Greece) also transferred
  - MegaPharm granted rights in Israel
- SOBI committed to order significant amount of additional vials
  - EUR 1.5 million over four quarters

US Commercialisation Strategy

- Commercialisation agreement with Santarus for North America
  - Small target audience of prescribers (~1000) to be covered by specialty field force of approximately 25
- Important potential milestones over coming 12-24 months
  - $10 million on successful read out of study 1310
  - $ 5 million on BLA acceptance
- SPA agreed with FDA in August
  - Provides clear regulatory guidance on way forward
- Follow-on indications being jointly developed/funded

Potential positioning: US (Acute)



Kalbitor:

- Black box warning on anaphylaxis on label
- Admin is difficult and storage not optimised
- Pharmacology explains level of reported efficacy

Berinert:

- Incomplete label (no peripheral and laryngeal attacks)

Firazyr

- Almost 100% (very) painful SC injection
- Relatively high level of estimated attack recurrence (32%)

Icatibant Clinical Briefing Document, CDER, FDA, 2011./ Aberer, et al. Ann Allergy Asthma Immunol 2010; 105(5):P238

Solidifying the HAE franchise

- Study C1113 Skin prick test (re- confirmation of clean immunological profile)
  - Q2, 12 completion
- Study C1209 EU Paediatric trial
  - Q4, 12 completion
- Study 1310 US Phase III (SPA agreement with FDA
  - Ongoing completion by 3Q/12 (triggers $ 10 million milestone)
- Study C1412 PASS (EU post approval patient registry)
- Home use label: programme in progress
- HAE prophylaxis: programme in preparation

Maximizing value rhC1 inhibitor franchise

- AMR Rationale
  - Circulating donor specific Abs leading to complement activation and cell death
  - Incidence < 5% of transplants
  - Add on therapy for one week during acute attacks
- AMR Status
  - Initiated Phase 2 study (PoC) including safety cohort
- AMR read out
  - Safety cohort expected Q1, 2012
  - PoC expected H2, 2012

Ischaemic reperfusion injury

- DGF rationale
  - Defined as need for dialysis in the immediate post transplant period
  - Incidence 20 to 30% of transplants (15 000/year in EU, similar in US)
  - Focus on “compromised grafts” (donor age, cold time, risk factors)
  - C1 effect through alternate and lectin pathways
- DGF status:
  - Pre-clinical study to refine pharmacodynamics in IRI underway
- Read out
  - Q4, 2011
  - Q1, 2012 next steps tbd

Validated transgenic expression platform

Expression and high yields (> 1 g/L) of recombinant human proteins in milk of transgenic animals

Granting of US patent (2027) further extends protection on core technology platform

- **Plasma Proteins**
  - Serpins: C1 Inhibitor, α1-antitrypsin and Antithrombin-III
  - Clotting Factors VII, VIII, IX and von Willebrand Factor
  - Albumin and Fibrinogen
- **Metabolic enzymes**
  - α–Glucosidase
- **Monoclonal antibodies**
  - High expression levels in various species: up to 30 g/L reported
- **Hormones**
  - Human Growth Hormone
- **Structural proteins**
  - Collagen
- **Others**
  - Protein vaccines
  - Lactoferrin and Lysozyme
  - Bile-salt stimulated lipase

Validated transgenic expression platform

- Our engine of value creation
  - Seeking collaborations
  - Strong IP
    - Granting of US patent (2027) further extends protection on core technology platform
- Relevant for most therapeutic proteins
  - Complex proteins of high quality with relatively high yields
  - Initial focus on blood clotting factors & metabolic enzymes
- Expression and high yields
  - (> 1 g/L) of recombinant human proteins in milk of transgenic animals

Financial highlights

- Revenues progression
  - Revenues & other income increased to €1.4 million (H1 2010: €0.1m)
- Continued focus on cost containment
  - Operating costs decreased to €8.8 million (H1 2010: € 10.1m)
  - Cash outflows of €8.9m (H1, 2010: €8.4m excluding DNage)
  - but 2010 period included significant (€ 3.0m) partnering cash income
- Significant reduction in net loss to €8.0 million (H1 2010: €28.0m)
  - Includes one-time €0.6m profit on discontinued operations (H1 2010: €1.7m loss)
- Cash at June 30, 2011 of €11.0 million (FY 2010: €10.5m)
  - Was strengthened further post period by private placement (€3.2m)

Financial Highlights

	Q2, 11	Q2, 10
Liquidity position (€M)	11.0*	9.8
Equity (€M)	4.1	11.4
Net cash used for operating activities (€M)	(8.9)	(10.1)
Operating Loss (€M)	(8.8)	(10.1)
Net loss (€M)	(8.0)^	(28.0)
Convertible debt (€M)	n/a	11.7
Number of shares outstanding	461,116,470**	304,953,323

*** Strengthened post period by private placement (€3.2m)**

^ Operating loss from continuing operations

*** * 490,116,470 following post period financing**

Upcoming Milestones

- Continue EU rollout of Ruconest
  - Ongoing
- Expand geographic coverage of Rhucin franchise
  - May 2011- Israel
  - ROW – Initiated significant nr of dialogues at BIO/ Follow up post BIO ongoing
- Expand rhC1-INH platform
  - AMR ongoing
    - safety readout Q1, 2012
    - PoC readout H2, 2012
  - IRI evaluation ongoing
- Leverage potential of the platform
  - Initiated significant nr of dialogues at BIO/ Follow up post BIO/ SE- Asia ongoing

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